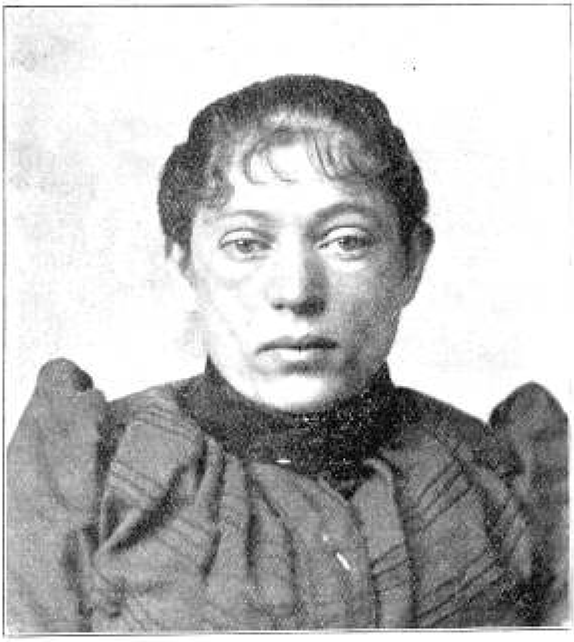
THE PHILADELPHIA NEUROLOGICAL SOCIETY.

November 27, 1899.

The President, Dr. James Hendrie Lloyd, in the chair.

A CASE OF MYOPATHY LIMITED TO THE FACE.

Dr. F. X. Dercum presented a woman, aged twenty-two; Russian; unmarried, whose father, aged sixty years, and mother, aged sixty-eight years, were both living and well.



She had one brother and one sister who were also perfectly well.

The patient had always been in good health up to six or seven years ago. At that time a "swelling" made its appearance in the back of the neck. The swelling suppurated and was lanced. It was attended by a great deal of pain. About this time or shortly after, she noticed that her cheeks were becoming thin and this atrophy steadily and slowly continued until the present.

Her cheeks are now much flattened and concave, due to wasting of the facial muscles. The orbiculares palpebrarum are much wasted as are also the zygomatics and the buccinators. The lips are large and full; the orbicularis oris seems to have suffered less than the other muscles of expression. The angles of the mouth are quite full and slightly depressed. The muscles of mastication are not involved. When the patient smiles, the angles of the mouth are drawn upward, but not outward, or very slightly so. During rest the orbicularis oris as a whole drops somewhat forward, suggesting, in a slight degree, the "tapir-like" mouth observed in more advanced stages of the disease. When the patient is told to forcibly close the eyes, the resistance of the lids can be readily overcome. The wasting of the orbiculares palpebrarum makes the eyes appear unusually prominent.

Electrical changes other than a slight quantitative diminution were not present.

A careful general examination of the patient was entirely negative. There was no involvement of the scapular, shoulder or arm muscles, nor of the legs or trunk. The case is interesting in the fact that the affection is limited to the face, differing in this respect from cases of facial myopathy hitherto described. In all the reported cases there appears to have been an involvement of other muscles, especially the scapular and humeral muscles constituting the so-called facio-scapulo-humeral type of Landouzy and Dejerine.

Cutaneous sensibility, organs of special sense, knee-jerks and visceral functions were normal.

A CASE WITH TABETIC SYMPTOMS AND EXAGGERATED KNEE-JERKS.

Dr. W. G. Spiller presented a white woman of forty-two years, who had been referred to his clinic from Dr. Hansell's clinic by Dr. Reber. She had color-blindness and her vision was much impaired. Syphilitic infection was possible. Within the past year she had had severe shooting pains in the extremities. She had also had temporary ptosis and diplopia. She was easily fatigued, and when fatigued, staggered. Disturbance of micturition had existed for years. Slight ataxia was detected when the gait and station were carefully tested. Both knee-jerks were exaggerated, more especially the left. The pupils were unequal and Argyll-Robertson sign was present. Dr. Reber had found that she had incipient optic atrophy with contracted fields. During the past year she had had "giving way of the legs." Babinski's reflex was not obtained.

Dr. Spiller thought that this case was either one of tabes or spinal syphilis, and that it might be an atypical form of the latter disease. The absence of the Babinski reflex indicated that the central motor tracts were not involved, and the exaggerated knee-jerks could not, therefore, be attributed to degeneration of these tracts if Babinski's sign were accepted. Optic atrophy was more characteristic of tabes than of syphilis. If the knee-jerks had been absent instead of exaggerated, the other symptoms were so marked that probably no one would have hesitated to call this a case of tabes.

Dr. F. X. Dercum said that he had had under observation a similar case in which there were pains, Argyll-Robertson pupil, well-preserved knee-jerks, and optic atrophy.

Dr. F. S. Pearce remarked that these cases were evidence of the fact that the sclerosis in its incipiency may be limited to the upper portions of the cord rather than the lower.

MELANCHOLY AND HYPOCHONDRIASIS—CURED BY GYNECOLOGIC OPERATIVE SUGGESTION.

Dr. F. Savary Pearce reported the following case:

R. F., American, single, aged twenty-five years, reported to the Howard Hospital Nervous Clinic, October 25, 1898. She complained of an imaginary ailment—pregnancy—and was extremely hypochondriacal and melancholy as to the dreaded outcome of her state. A sadder picture of a delusion he had seldom seen. There was no history of hereditary nervous disease. She began to menstruate at fourteen years and suffered somewhat from the beginning with dysmenorrhea. Her mother died when the patient was twenty years old. This depressed her very much. Then she had added household duties, and this probably with a run-down condition added to the gradually increasing dysmenorrhea. In August, 1898, she imagined she had been raped and become pregnant, but it is almost certain that coitus never took place. The next menstrual period not coming until three days late, the woman was positive of pregnancy and had heard so much on the subject from an aunt that her salvation seemed to be the taking an oxytocic which she did in the shape of several cupfuls of a strong decoction of pennyroyal. This is said to have established the flow precipitously and with much pain. It should be mentioned that she had vomited the day before taking the drug and that this clinched the idea of pregnancy. Since then the periods have occurred about every three weeks. When she visited the clinic she was convinced that the drug had not been effective, and that she was still pregnant.

When examined on October 25, 1898, she was melancholic and hypochondriacal, extremely emaciated and delusional as to pregnancy. She gave no evidences of other insanity, the one idea dominating all else was the rape and pregnancy. Tonics were employed for the building up of her depraved nervous system. A persistent acne and anemia were somewhat improved bv Blaud's pill and hygienic measures, but she could be induced to take but little food and her mental state remain unchanged. Finally on March 6, 1890. Dr. John B. Shober found the symptoms of obstructive dysmenorrhea. Pelvic examination showed acute anteflexion of uterus with elongation of supravaginal cervix—otherwise normal. Hymen absent. Dilatation of cervix and curettement of uterus were done under ether. The uterine sound entered four inches. A cure by this operation was promised.

The patient remained in the Gynecean Hospital several weeks under Dr. Shober's care, and when discharged she seemed more composed and her mental condition was greatly improved. On May 20, 1899, the note was entered that she was gaining flesh rapidly and had improved in every way, the delusion of pregnancy having remained away. Her menstrual periods had returned and were now practically normal, perhaps somewhat too frequent.

While there was slight gynecologic disease in this case sufficient to set up irritation, as shown by painful menstruation, still the cure of the mental depression seemed to be entirely attributable to the suggestion of doing away with the imaginary impregnation by the curettement.

EPILEPTIC AMBULATORY AUTOMATISM.

INTESTINAL AUTOINTOXICATION PROBABLE EXCITER OF SOME OF THE ATTACKS.

Dr. F. S. Pearce reported also the following case: A. F. G., aged 28 years, male, a very intelligent journalist, had enjoyed good health. He had always been a hard mental worker. Five years ago he had a very severe attack of septic typhoid fever with relapses; followed by post-typhoid meningitis. Convalescence was very long. For two years he was treated for persistent gastro-intestinal catarrh, followed a year later by marked gastrectasia and gastroptosis. Three years ago he had much improved in general vigor and somatic symptoms after a most rigid diet consisting largely of milk. His "stomach always remained sensitive," and from slight indiscretions in quality or excess of food he would suffer much for a fortnight from gastro-intestinal fermentation, depression, insomnia and palpitation of heart. He remained anemic. One day, from no apparent cause, he fell over after supper in a typical attack of grand mal. He has had half a dozen such attacks during the past three years, always preceded by especial exacerbation of his chronic dyspepsia. During the past eighteen months he has had spells of acute indigestion, belching of wind, and tympanites. With these he first developed the epileptic phenomenon to be described, an attack of which was seen by Dr. Pearce on July 25, 1899, and which was as follows: The attack began by the patient calling for his wife. He was found running about the hall and into the bathroom in a dazed condition. He had the peculiar epileptic fixity of eyes and was much confused. He would talk of things that occurred during the day, finally stopping his wanderings to grasp tightly the bedstead, and belching a large amount of gas with some acrid stomach contents. He then was able to walk to his bed with assistance. He had no convulsion whatever, and from a dazed condition fell into a profound post-epileptic sleep from which he awoke in half an hour quite restored. A sea bath later in the afternoon caused him to belch gas freely.

The dilated stomach and evident state of intestinal fermentation in this man (indican was found in large quantities) led Dr. Pearce to the conclusion that primary intestinal irritation was the cause of the epileptic paroxysm, in a system predisposed to the epileptic state most likely by the post-typhoid meningitis referred to above. The sequence of indigestion from slight cause, discomfort in the epigastric region, borborygmus, and acrid eructations, has been too frequently seen to permit a doubt that such irritation does induce attacks at times as in the case recorded. The use of antifermentative treatment (salol) and the cutting down of the bromide of strontium to gr. x, *t. i. d.*, have much improved his general health, and the attacks have been less severe and considerably reduced in number.

A CASE OF SYPHILITIC PARAPLEGIA OF THE ARMS, THE LEGS ESCAPING.

Dr. F. X. Dercum presented a colored man, aged 29 years, who said that he had a chancroid (?) about four years ago. About seventeen months ago he was unable to move the right eyeball on account of paralysis of the muscles, or to raise the eyelid. He had no loss of vision, but suffered greatly from pain on the top of the head. In about three weeks both the paralysis of the eye and the headache disappeared.

About fourteen months ago the fingers of the right hand became weak and he could not properly open them or hold anything in them. Later the arm became weak and he could not raise it over his head. This condition came on gradually. About six months after the appearance of the symptoms on the right side the same condition occurred on the left, and very soon, the patient states, he could not raise either of his arms above his head. The right hand slightly improved with time, while the left became somewhat worse. He occasionally had cramps in the muscles of his arms and fingers. He noticed that when the fingers were closed on the palms, he could not open them. The onset of the paralysis was accompanied by pain which was sharp and shooting in character, and worse at night. The pain seemed to begin in the neck and spread down the arms.

The headache persisted with varying intensity, and was also worse at night. For a time he was troubled with sleepiness, though for three weeks past he had suffered from insomnia.

There was no history of secondary syphilis. He did not have any internal treatment at any time.

At present the patient presents a striking and marked paralysis of both arms. He has paresis of both deltoids, and is unable to raise the arms beyond a right angle with the body. This is equally marked on both sides. He has also marked palsy of both triceps muscles and of the extensor groups of both forearms. In addition there is present marked wrist-drop in the left hand and almost complete wrist-drop of the right hand. He is also unable to extend the fingers; these assume the position of flexion due to the unopposed action of the flexors. The palsy of the extensors of fingers is most marked in the left hand, where it is complete. Neither the thumb nor any of the fingers can be in the least degree extended. In the right hand he has still slight power of moving the fingers and thumb in extension. Both biceps muscles, though small, are strong and show no evidences whatever of palsy.

In the arms there is a very pronounced wasting of both the flexor and extensor groups. In the left forearm the extensor group especially is markedly flattened. The supinator longus muscle, on the other hand, is well preserved, supination being performed readily and in an apparently normal manner. The interosseous muscles are decidedly wasted. There is no apparent wasting of the thenar and hypothenar eminences, although these muscles are exceedingly soft. In the right forearm there is likewise a flattening of the flexor and extensor groups, though the wasting of the flexor group is much less marked than in the left arm. Wasting of interosseous muscles in right hand, while evident, is relatively slight. The thenar and hypothenar eminences are normal. The supinator longus muscle as in the left arm is well preserved. Pronation and supination are performed with equal readiness in both forearms.

Electrical examination reveals marked quantitative diminution in the wasted muscles.

The movements of head and neck are performed freely in all directions. He states that there is a feeling of cramp or stiffness when bending the neck backwards.

There is no involvement of the legs. They are not paretic, nor is there any change in the gait. There is no involvement of the bladder, nor is there any history of such involvement. The tendon reactions are normal. This is true equally of the elbow-jerks and knee-jerks. The plantar reflexes also are normal. Sensation is well preserved to all forms. Pupils are unequal, the right being the larger; both react to light and in accommodation. There is a ptosis of the right lid; also paresis of the left externus.

The case is interesting because it departs so widely from the common form of spinal syphilis, the form which has been described by Erb. Cases of syphilis of the cervical cord without symptoms of lumbar involvement are extremely rare, and in the writer's experience the present case is unique. It is interesting further because of the almost symmetrical involvement of the arms; the paralysis is most marked in the same muscles and groups of muscles; for instance, in the triceps and extensors of the forearm upon both sides, while, on the other hand, the biceps and the long supinator upon both sides entirely escape. It is important further to note that the palsy to some extent approximates the form of brachial palsy known as the lower arm type, and this fact enables us to localize the lesion with considerable accuracy. The lesion must involve the cord slightly above or at the level of the seventh and eighth cervical and first dorsal roots. That it was not absolutely confined to these levels is proven by the partial involvement of the deltoids, though the remarkable preservation of the biceps and supinator longus shows that the lesions at the upper brachial levels cannot be pronounced.

Another point worthy of attention is the fact that the involvement of the arms, though symmetrical, is not equal in degree, one arm being more decidedly affected than the other; we have here an analogy with syphilis of the lumbar cord, the involvement being generally a little more pronounced upon one side than upon the other.

Note—Since the patient was presented to the society active specific treatment has been instituted, with the result of a very marked improvement in the paralysis. The patient is now able to perform all of the normal movements of the arms, while all of the paralyzed muscles show a decided return of strength. This observation is important as demonstrating the undoubted syphilitic nature of the case.

ISOLATED ABDUCENS PALSY, PROBABLY PERIPHERAL.

Dr. Wendell Reber, by invitation, reported a case of isolated palsy of the left external rectus. A woman of 50 years came to the clinic of Dr. Hansell at the Polyclinic Hospital with paralysis of the left external rectus. Four weeks previously she awoke with rather obscure vision and more or less diplopia. These conditions became more pronounced the following day. No history of trauma or of any antecedent acute disorder except rheumatism could be obtained. Two brothers and two sisters of the patient were the subjects of a more or less aggravated lithemic middle ear deafness, and the patient herself was deaf. No nervous or orbital disease could be found to explain the abducens palsy. The woman slept with the head of her bed between two windows, and with much draught upon her. She also slept on her right side, so that the left side of her face was exposed. The case was thought to be one of palsy in a rheumatic person following exposure to draught. Considerable motion was regained in paralyzed muscle by the use of strychnine, the iodides, the salicylates, electricity, etc.

Dr. Reber said that little could be found in the literature concerning such cases, and quoted the opinions of several writers.

Dr. William G. Spiller said that he had been much interested in this case because of the possibility of the abducens palsy being due to rheumatism, or exposure to draught; as occurs in facial palsy. The patient slept with the affected side of the face exposed to a draught; she had had rheumatism two years ago, and at one time had facial palsy. About a year ago Dr. de Schweinitz read a paper before the society reporting two cases of retrobulbar neuritis in patients who had had facial palsy. A relation seemed to exist between the two conditions. Dr. Spiller thought that abducens palsy might be related to facial palsy in the same way, as he had a case under observation, sent to his clinic by Dr. Risley, in which abducens palsy followed facial palsy of the same side a day or two after the exposure to a draught.

A CASE OF AMYOTROPHIC LATERAL SCLEROSIS IN WHICH DEGENERATION WAS TRACED FROM THE CEREBRAL CORTEX TO THE MUSCLES.[1]

WITH REMARKS ON THE EXTENT OF THE CORTICAL MOTOR AREA IN MAN.

This was the title of a paper read in abstract by Dr. Spiller. Dr. Spiller said that the cases of amyotrophic lateral sclerosis with necropsy and microscopical examination are not very numerous, and those in which the degeneration of the central motor tracts has been traced throughout the cord and brain into the central cortex number only ten, and in only two of these (case of Probst and case of Spiller) has the corpus callosum been examined and found diseased.

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[1] This paper will be published in full in the memorial volume to be issued from the William Pepper Laboratory of Clinical Medicine.

In the case reported by Dr. Spiller the degenerated motor fibers were traced throughout the central nervous system from the Rolandic area to the sacral region, and degeneration was found in the corpus callosum, ulnar nerve, and the muscles of an upper limb. The degeneration of the cerebral cortex was employed in the determination of the extent of the cortical motor area in man.

The patient was in the service of Dr. C. K. Mills at the Philadelphia Hospital, and the clinical notes and pathological material were obtained from Dr. Mills. A brief abstract of the case was as follows:

A man, 55 years of age, a laborer, contracted a severe cold in October or November, 1897. He said he had been healthy until that time. Toward the end of 1897 he began to have dysphagia, involuntary dribbling of saliva and loss of power in the legs and arms. These symptoms increased until he became confined to his bed, and was almost completely paralyzed. The deep reflexes were exaggerated, and fibrillary tremors were noticed in the tongue and other muscles. Sensation was not disturbed. Muscular atrophy was intense. Death occurred November 17, 1898.

The disease began with bulbar symptoms and involvement of the limbs, and ran a rapid course, the patient dying about a year after his symptoms first became very perceptible.

The muscles of the upper extremity examined were atrophied, and those near the distal end of the limb were more altered than those near the proximal end. In an interosseous muscle examined the atrophy was found in an extreme degree, the muscular fibers were small, but many still preserved the transverse striation; some had only longitudinal striation. Fatty degeneration of muscular fibers could not be found. The interstitial connective tissue was increased in amount, and much interstitial fatty tissue was found. The intramuscular nerve fibers and the muscle spindles appeared to be normal.

The ulnar nerve when teased and stained with osmic acid was found to be partly degenerated. The changes in the ulnar nerve were comparatively slight.

The anterior roots of the spinal cord were much atrophied, and in some sclerotic areas were found.

The nerve cells of the anterior horns were greatly degenerated, especially in the cervical region, although those in the lumbar region were also much diseased.

The crossed pyramidal tracts and the right direct pyramidal tract were degenerated throughout the cord from the motor decussation into the lumbar region. The sclerosis of the crossed pyramidal tracts could be followed into the sacral region. Degeneration in the motor tracts could be traced by Marchi's method from the lumbar region to the motor cortex.

The hypoglossal nuclei and intramedullary portion of the hypoglossal nerves were altered. The posterior nucleus of the vagus contained some highly pigmented cells. The nucleus ambiguus contained few cells, and some of these were possibly diseased. The seventh nuclei were normal, except that many of the cells were much pigmented. The posterior longitudinal bundles were slightly altered.

Distinct degeneration of the fibers within the motor area of the cortex was found by Marchi's method, and the degeneration of the corpus callosum was intense. The cortical nerve cells within and without the motor areas were greatly pigmented, and black granules were found by Marchi's method in the area of the tangential fibers. The giant cells of the paracentral lobule were numerous, but much pigmented.

The cases of amyotrophic lateral sclerosis in which degeneration has been traced throughout the pyramidal tract to the cortex are two of Kojewnikoff, two of Charcot and Marie, one of Lennmalm, one of Lumbroso, one of Hoche, one of Probst, one of Mott, and one of Spiller, numbering ten in all literature.

The degeneration of the corpus callosum was intense in Dr. Spiller's case. Distinct parallel rows of black dots could be seen all through the sections from the middle portions of the callosum. The connection of the motor areas of the brain as indicated by these findings must be a very intimate one.

The changes in the peripheral muscles and ulnar nerve in Dr. Spiller's case were interesting. Those muscles nearest the distal end of the upper limb were most wasted. The greatest alteration was found in the first interosseous, and the supinator longus was almost as much diseased, while the biceps and deltoid were not so much affected. Even in the first interosseous, normal muscular fibers were found in the midst of the intensely atrophied ones. In the supinator longus atrophied fibers were found, occurring in groups in close proximity to normal ones. This explains why reaction of degeneration may be absent in amyotrophic lateral sclerosis. The microscopical study shows that one muscular fiber after another undergoes almost complete atrophy, and that even late in the process normal muscular fibers are found in the most altered tissue.

In these highly atrophied muscles the intramuscular nerves were normal. There is no reason for the belief that atrophy of muscles of necessity causes alteration of intramuscular nerve fibers, detectable by the Weigert's hematoxylin method at least. Probably some of these intramuscular nerves were sensory.

The muscular fibers in sections taken from the right side of the tip of the tongue were much atrophied, but by Marchi's method did not contain any evidence of fat. Many muscular fibers were perfectly normal. In view of the distinct atrophy in the twelfth nuclei, and of the fibrillary tremors of the tongue seen during life, this alteration of the lingual muscles was expected. The interstitial fatty tissue was very abundant, and many of the muscular fibers were very small. The intramuscular nerve fibers of the tongue stained by Weigert's hematoxylin method appeared to be somewhat degenerated.

Amyotrophic lateral sclerosis is essentially a disease of the motor system, and it occurred to Dr. Spiller that the degeneration of the cerebral cortex in his case could be employed in determining the extent of the cortical motor area in man. This has never been done in any of the few cases of amyotrophic lateral sclerosis with cortical degeneration reported in the literature.

Our knowledge of the extent of the motor cortex in man is very incomplete. Lemacq has attempted to define the motor area by the results of the electrical irritation of the human brain at the time of operation, and he has depended very largely on the work of American investigators (Keen, Mills, Starr, Dana, Lloyd, and others). The diagram of the motor area determined in this manner is imperfect.

Dr. Charles K. Mills said that the paper read by Dr. Spiller was one of the most valuable contributions ever presented to the society. It demonstrated the extent of the motor cortex in man by an entirely original method, and the pathological findings in general were of unusual interest and importance. It seemed to him that Dr. Spiller's results, in so far as they fixed the limits of the motor cortex, were a further confirmation of the views which the speaker had long held and taught, viz., that the motor region is in all essential respects separate from the sensory region of the cortex. This motor area he believed is limited, as shown in his published diagrams, almost exactly as determined by Dr. Spiller. Dr. Mills believed that the central convolutions and the parts corresponding to them on the median surface of the hemispheres, and the posterior extremities of the first and second, and, to a certain extent, of the third frontal convolution, together constitute the cortical motor area. He also still held that the great sensory area of the cortex (the region of representation of muscular and cutaneous sensibility), is composed of the superior parietal lobule, in part of the inferior parietal lobule, of the quadrate lobule and the gyrus fornicatus.

In a strictly practical field he had recently had a further confirmation of these views. One of his patients at first had only ataxia and loss of the stereognostic sense, with impairment of all forms of cutaneous sensibility. Later there was added paresis and finally paralysis of the arm and leg and slight paralysis of the face, with some wasting, verbal amnesia, and temporary visual symptoms. In this case Dr. Keen had successfully removed a cystic tumor, which was mainly located in the superior parietal lobule, although it had grown somewhat forward and inward from this position. The diagnosis of a tumor in the superior parietal lobule had been made two months before the date of operation because of the ataxia and the sensory symptoms.

Dr. D. J. McCarthy asked Dr. Spiller whether the failure to trace the degeneration from the crossed pyramidal tracts into the internal capsule in a case of amyotrophic lateral sclerosis reported by Dercum and Spiller, was because the material was exhausted, or because there was no degeneration present. Also in regard to the presence of degeneration in the posterior longitudinal bundles. He also inquired whether Dr. Spiller considered the degeneration in the antero-lateral column a primary degeneration of motor fibers.

Dr. E. L. Mellus inquired in regard to the degeneration in the corpus callosum, and also whether it was possible to make out any grouping of motor focal points as suggested by Bevin Lewis' researches, and possibly by the fact that under electrical stimulation the motor points in man seem to be more separate than in the lower animals?

Dr. Spiller said that the posterior longitudinal bundles were slightly degenerated. The reason the degeneration was not traced above the pons in the case reported by Dr. Dercum and himself was because the degeneration ceased at this point and not because of lack of material. A similar condition has been observed in cases of paretic dementia. The degeneration in the corpus callosum was most intense in the middle portion of the part that is seen when the hemispheres are pressed aside. There was some degeneration in the knee of the corpus callosum, but degeneration in the splenium was not distinct.

With regard to the grouping of motor focal points within the motor cortex, as mentioned by Dr. Mellus, Dr. Spiller said he could not give any information. He had not been able to observe any difference in the degree of degeneration in sections taken from the same block.

With regard to the antero-lateral column, Dr. Spiller said that degeneration of this portion of the cord is not a very uncommon finding in amyotrophic lateral sclerosis. This degeneration may be in fibers connecting the cells of the anterior horns of different levels with one another.

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21. EIN FALL VON DIFFUSER SARKOMATOSE DER GESAMTEN PIA MATER DES GEHIRNS UND RÜCKENMARKS (A Case of Diffuse Sarcomatosis of the Entire Pia of the Brain and Cord). Schröder (Monatsschrift für Psychiatrie und Neurologie, 6, 1899, p. 352).

The symptoms in this case were vomiting, choked disks, headache in the occipital and parietal regions, uncertainty of gait, exophthalmos, Romberg's sign, left facial paresis, deviation of the tongue to the right, diminution of the knee-jerk on one side and absence on the opposite, partial deafness, hallucinations of touch, etc. The cause of these symptoms was sarcomatosis of the entire pia of cord and brain, extending into the substance of the brain, cord, cerebral nerves and spinal roots. A brief review of the few cases of sarcomatosis of the central nervous system reported in the literature is given. SPILLER.